

1 **Lung transplantation for Kartagener syndrome: technical aspects and morphological**
2 **adaptation of the transplanted lungs**

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22 **Key words:** Kartagener syndrome, lung transplantation, morphology, technique, surgery

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1 **Abbreviations**

2 **CT**, computed tomography

3 **CPB**, cardiopulmonary bypass

4 **KS**, Kartagener syndrome

5 **LT**, lung transplantation

6 **PA**, pulmonary artery

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1 **Abstract**

2 While technical considerations in lung transplantation for Kartagener syndrome have been
3 discussed, little information is available about the postoperative morphological changes of the
4 grafted lungs. Herein, we discuss both the technical aspects and postoperative morphological
5 adaptation of the grafted lungs in a case of Kartagener syndrome. A 46-year-old male patient with
6 Kartagener syndrome underwent bilateral cadaveric lung transplantation. The right arterial
7 anastomosis for transplantation of the size-matched grafts required technical elaboration. After
8 the transplantation, we found a free space in the cardiac notch of the left lung and partial collapse
9 of the lower lobe of the right lung due to dextrocardia. Follow-up computed tomography performed
10 on day 42 after the transplantation demonstrated resolution of the atelectasis and morphological
11 adaptation of the grafts into the recipient's chest cavity with dextrocardia. Considering such early
12 morphological adaptation of size-matched grafts, lobar reduction could be avoided in lung
13 transplantation for Kartagener syndrome.

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1 **Introduction**

2 Kartagener syndrome (KS) is a rare congenital disorder associated with primary ciliary dyskinesia,
3 that is clinically characterized by the triad of situs inversus totalis, bronchiectasis, and chronic
4 sinusitis. Lung transplantation (LT) has been demonstrated as a feasible therapeutic option for
5 patients with KS who present with progressive bronchiectasis and chronic respiratory failure, with
6 similar reported long-term outcomes to those of LT for other diseases [1-6]. In LT for KS, an
7 elaborate surgical technique is required to adjust for the orientation mismatch of the hilar
8 structures between the recipient and the donor, especially for pulmonary arterial anastomosis, as
9 applied in inverted LT [1-7]. Moreover, the normal anatomical lungs from the donor is
10 morphologically necessitated to fit into the recipient's chest cavity in the presence of situs inversus
11 with dextrocardia. Morphological mismatch between the donor normal lung and the recipient's
12 dextrocardia might limit full expansion of the right transplanted lung. As a solution to this
13 morphological mismatch, right lower lobectomy of the donor lungs has been performed previously
14 [1, 3]. By contrast, recent reports have shown the feasibility of LT without lobar reduction in
15 patients with KS [2, 4, 5]. However, the postoperative morphological changes of the transplanted
16 whole lungs in patients with KS remains unknown. The observed morphological adaptation of the
17 transplanted lungs to the recipient's chest cavity with dextrocardia in the early phase after LT
18 might validate the avoidance in LT for KS of lobar reduction, which is associated with a risk of
19 bronchial fistula formation [3]. Herein, we report a case of bilateral cadaveric LT for a patient with
20 KS, describing the technical aspects of the operation and the postoperative morphological
21 changes of the transplanted whole lungs, which were assessed by computed tomography (CT).

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23 **Case**

24 A 46-year-old male patient with KS and adrenocorticotrophic hormone deficiency required long-
25 term supplemental oxygen therapy and macrolide therapy due to recurrent lung infections for
26 more than 3 years, and was considered for bilateral LT. Chest X-ray and CT showed

1 bronchiectasis and pulmonary cystic changes predominantly in the right lung, as well as situs
2 inversus totalis (**Figure 1**). The patient had undergone planned sinus surgery and emergent
3 bronchial artery embolization for hemoptysis during the waiting period for LT.

4 Seventeen months after he was entered on the list, the patient underwent bilateral LT with
5 extended-criteria donor lungs from a 30-year-old male donor who died of non-traumatic cerebral
6 hemorrhage. The donor had a current smoking history with 14 pack-years and an arterial oxygen
7 partial pressure/fraction of inspired oxygen ratio of 262 due to mild atelectasis of the bilateral
8 lower lobes. The estimated vital capacity-based size match of the donor lungs to the recipient
9 lungs was 104%. Considering the severe condition of the recipient and scarcity of organ donors
10 in Japan, we decided to use the extended-criteria donor lungs. The donor lungs were fully
11 expanded with recruitment maneuvers and had no edema with an arterial oxygen partial
12 pressure/fraction of inspired oxygen ratio of 292 during the lung procurement. After the lung
13 procurement, the length of the donor pulmonary artery (PA) was preserved to facilitate the arterial
14 anastomoses during the back-table preparation of the double-lung block.

15 The operation was performed through a clamshell thoracotomy. Bronchial blocker
16 placement via a single-lumen endotracheal tube facilitated the subsequent bilateral bronchial
17 anastomoses. Severe pleural adhesions, especially in the right lung apex, necessitated careful
18 adhesiolysis before the establishment of cardiopulmonary bypass (CPB). CPB was established
19 to obtain cardiac drainage and to create a surgical field where the central hilar structures were
20 easily accessible. After bilateral pneumonectomy, the donor left lung was first transplanted in the
21 usual manner. For the left bronchial anastomosis, the short left main bronchus of the recipient
22 was readily anastomosed to the normal left main bronchus of the donor, despite the size
23 discrepancy. For the right lung transplantation, the donor atrial cuff necessitated suture
24 adjustment due to the size discrepancy in the right atrial anastomosis after the right bronchial
25 anastomosis. The right, but anatomically left PA, of the recipient running dorsally in the right chest
26 was dissected as proximally as possible for adequate mobilization. Subsequently, the right PA

1 proximal to the first branch of the recipient was divided leaving a beveled edge, with sparing of
2 the dorsal PA wall for orientation matching, facilitating end-to-end anastomosis (**Figure 2**).

3 After transplantation of both the lungs, we found a free space in the cardiac notch of the left
4 lung, as well as partial collapse of the right lower lobe due to the dextrocardia; however, we
5 decided not to perform lobar reduction, because we considered that the size matching was
6 adequate (**Figure 3A**). Neither kinking of the pulmonary vessels, nor congestion of the
7 transplanted lungs was observed after the reperfusion. Chest closure was performed without the
8 use of the artificial chest wall. The CPB time, operative time, and total ischemic time were 237
9 min, 648 min, and 603 min, respectively. The intraoperative blood loss was 3220 mL. No
10 postoperative anticoagulation was required.

11 Chest CT performed on day 2 after the LT showed pleural effusion in the cardiac notch of
12 the left lung and right lower lobe atelectasis (**Figure 3B**). By contrast, the follow-up CT performed
13 on day 42 after the LT demonstrated that the cardiac enlargement had resolved and that the
14 transplanted lungs had expanded to fit into the recipient's chest cavity with dextrocardia, with
15 filling of the space in the cardiac notch by the left lung and resolution of the atelectasis of the right
16 lower lobe (**Figure 3C**). The patient had an uneventful postoperative course and was discharged
17 from the hospital on day 65 after the LT. Postoperative lung perfusion scintigraphy revealed
18 homogeneous distribution of the blood flow in the transplanted lungs and the right-to-left ratio was
19 33 to 67. Unfortunately, the patient died of pneumonia 364 days after the surgery. However, no
20 radiological or physiological evidence of anastomotic problems was seen at any time-point during
21 the postoperative course.

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23 **Discussion**

24 To date, LT for KS has mainly been discussed from the point of view of technical considerations
25 on account of the situs inversus. To the best of our knowledge, this is the first report to describe,
26 in addition to the technical aspects, the morphological changes of the transplanted lungs, as

1 assessed by CT, after bilateral LT for KS. According to the CT assessment, the transplanted lungs
2 filled into the shape of the recipient's chest cavity with dextrocardia within 1.5 months, allowing
3 an uneventful postoperative course of the patient.

4 In LT for KS, the major concern is the technical aspects, especially in relation to the PA
5 anastomosis. Because of the risk of kinking of the right PA anastomosis after LT in patients with
6 KS due to the situs inversus in the recipient [2], technical modifications for anatomical variations
7 have been discussed in previous reports [4, 5]. In our case, to prevent kinking, we dissected the
8 right PA of the recipient proximally for adequate mobilization and divided the recipient PA leaving
9 a beveled edge, with sparing of the dorsal wall of the PA at the proximal level of the first branch
10 for subsequent PA anastomosis.

11 Moreover, the orientation and size mismatch between the donor normal lung and the
12 recipient's chest cavity with dextrocardia should be resolved in cases of KS undergoing LT. Since
13 the space in the right chest cavity in a patient with dextrocardia is limited, right lower lobectomy
14 was performed previously for volume reduction in cases KS undergoing LT [1, 3], which increased
15 the risk of bronchial fistula formation [3]. However, in the case reported herein, we used size-
16 matched donor lungs that allowed transplantation of the whole lungs without lobar reduction. CT
17 assessment demonstrated temporary atelectasis of the right lower lobe early after LT, but by 6
18 weeks after the LT, the atelectasis of the right lower lobe had resolved and the transplanted lung
19 had expanded to fill the space in the cardiac notch of the left lung that had been observed in the
20 early postoperative CT. Given this early morphological adaptation of the size-matched lung grafts,
21 unexpected lobar reduction might be avoided in LT for KS.

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23 **Conclusion**

24 Although the PA anastomosis requires technical elaboration, bilateral LT appears to be a feasible
25 therapeutic option for patients with KS. The observed early morphological adaptation of the size-
26 matched donor lungs into the recipient's chest cavity in the presence of dextrocardia might

1 suggest that lobar reduction could well be avoided in cases of LT for KS.

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1 **Compliance with ethical standards**

2 Informed consent: Written informed consent was obtained from the patient for publication of this
3 case report and any accompanying images.

4 **Conflicts of interest:** Haruchika Yamamoto and other co-authors have no conflict of interest.

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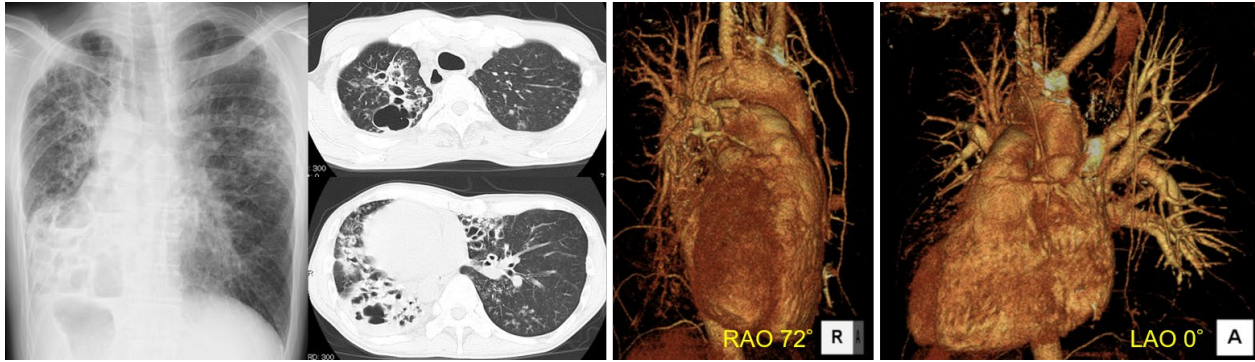
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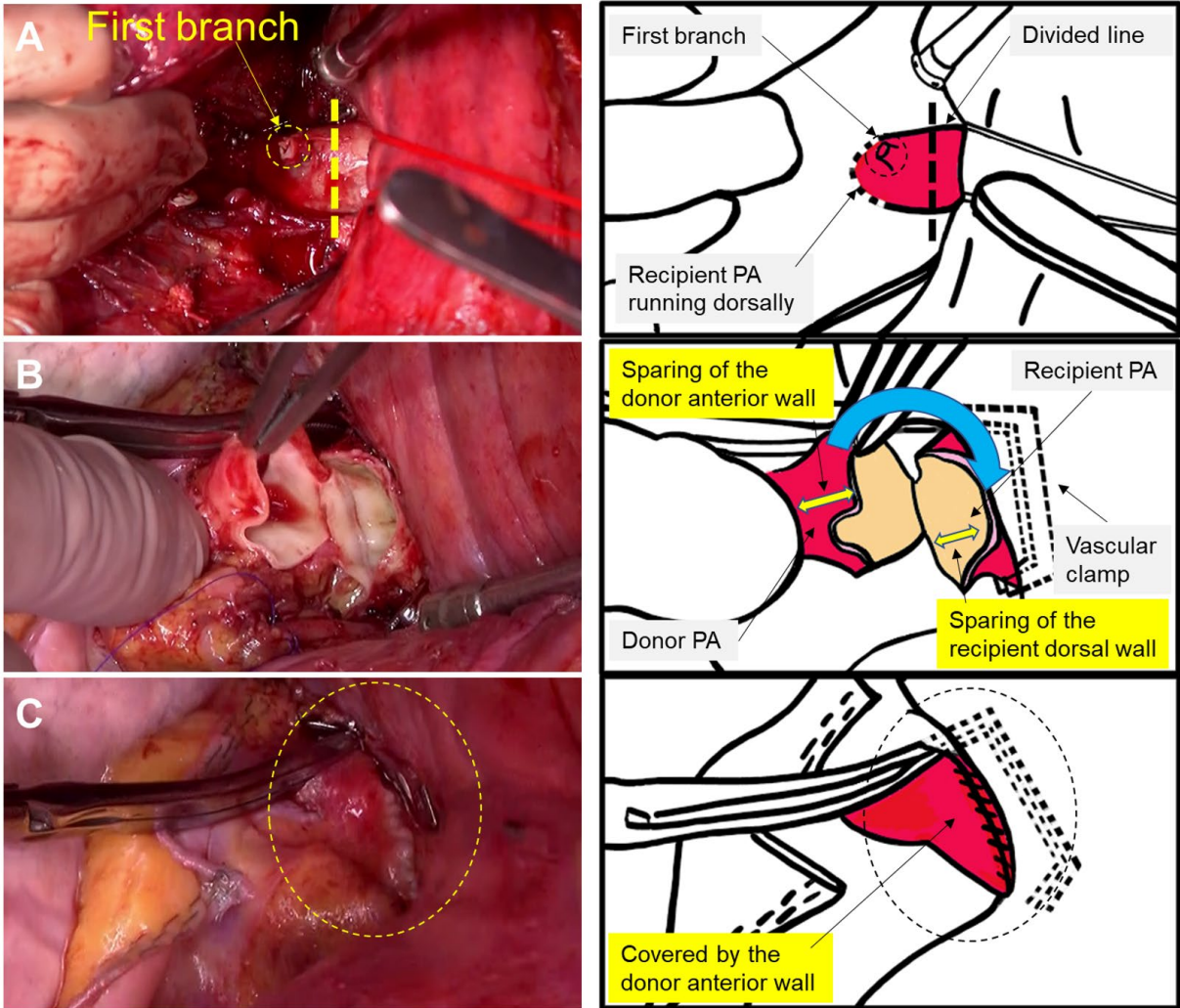
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1 **Figure legends**

2 **Figure 1.** Chest X-ray and computed tomographic images obtained prior to the lung
3 transplantation showing evidence of bronchiectasis and pulmonary cystic changes, predominantly
4 in the right lung, with situs inversus totalis.

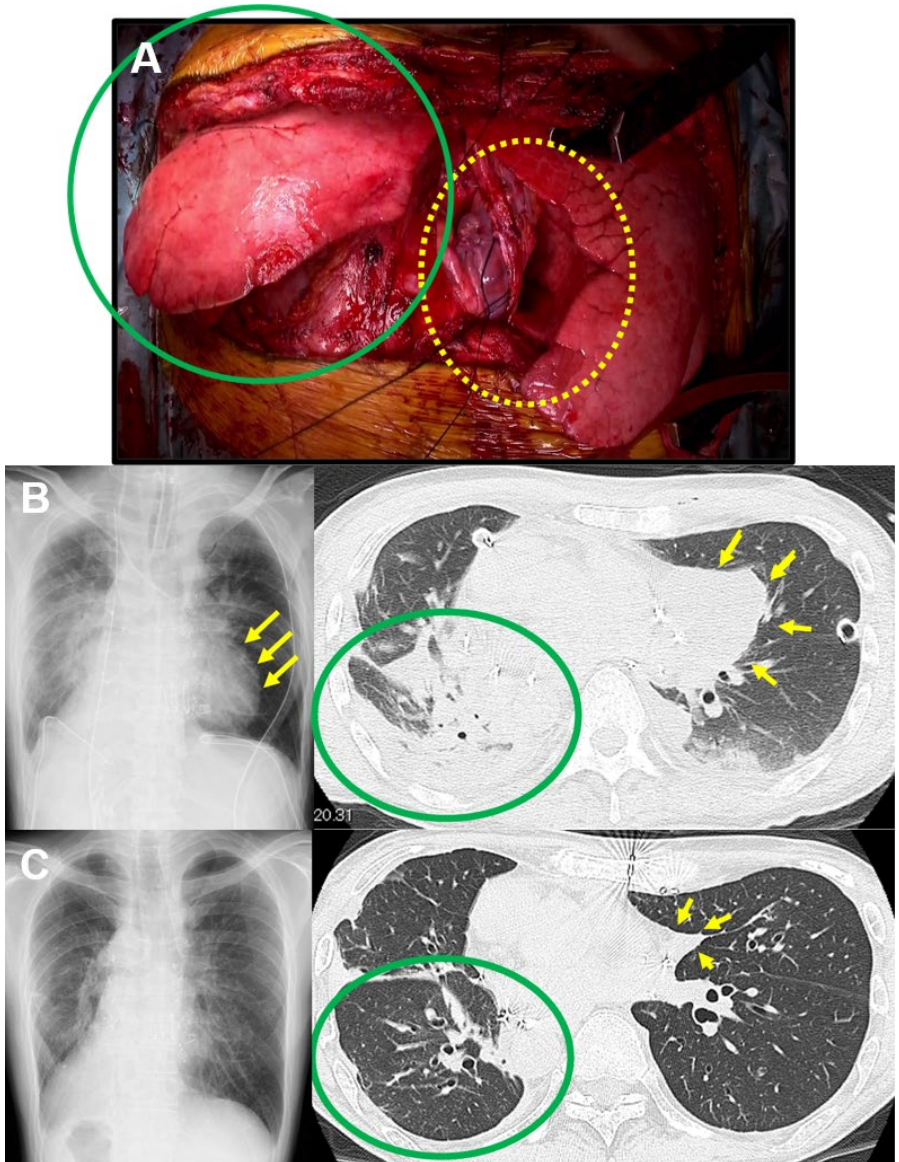


1 **Figure 2.** Intraoperative findings of the right arterial anastomosis. To prevent kinking, the right
2 main pulmonary artery (PA) of the recipient running dorsally in the thorax was dissected proximally
3 for adequate mobilization. The right main PA proximal to the first branch of the recipient (dashed
4 line) **(A)** was divided leaving a beveled edge, with sparing of the dorsal wall of the PA for
5 orientation matching **(B)**, facilitating subsequent arterial anastomosis (dashed-line circle) **(C)**.



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1 **Figure 3. (A)** Intraoperative findings before closure of the chest revealed the right expanded lung
2 in the small chest cavity with dextrocardia (green circle) and a free space in the cardiac notch of
3 the left lung (yellow dashed-line circle). **(B)** Chest X-ray and computed tomographic images
4 obtained on day 2 after lung transplantation showing atelectasis of the right lower lobe (green
5 circle) and pleural effusion in the cardiac notch of the left lung (yellow arrow). **(C)** Chest X-ray and
6 computed tomographic images obtained on day 42 after the lung transplantation showing
7 resolution of the cardiac enlargement and expansion of the atelectatic right lower lobe (green
8 circle), and expansion of the transplanted lungs to fill the space in the cardiac notch (yellow arrow).



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